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JUVENILE DERMATOFIBROSARCOMA PROTUBERANS OF THE SCALP: THERAPEUTIC MANAGEMENT CHALLENGES

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ABSTRACT :

Probably genetic in origin, dermatofibrosarcoma protuberans (DFSP) constitutes the most common type of fibrohistiocytic sarcoma. Classified as intermediate malignancy, this tumor is neither strictly malignant, as it does not metastasize, nor strictly benign, due to its high recurrence rate if not completely excised.

Therefore, its prognosis depends primarily on the quality of its surgical excision and especially on its histological analysis. We report an unusual presentation of dermatofibrosarcoma protuberans, characterized by its rare location on the scalp, distinctive clinical appearance in a 13-year-old girl. Subsequently, we will discuss our diagnostic approach in light of the literature and our surgical therapeutic strategy.

Keywords: Dermatofibrosarcoma protuberans, scalp, child, skin expansion.

INTRODUCTION:

Dermatofibrosarcoma protuberans (DFSP), is a superficial sarcoma with fibroblastic differentiation of the dermis and underlying soft tissue. It is characterized by a very slow progression and carries a significant risk of recurrence if not adequately excised.

This rare fibrous skin tumor represents 0.1% of malignant skin tumors and 2-6% of all soft tissue sarcomas [1]. According to the literature, the scalp is not commonly cited as an elective site for this lesion.

We report our experience in the diagnostic and therapeutic management of an atypical presentation of this tumor in a 13-year-old girl, occurring with a rare localization on the scalp and an unusual clinical presentation. We will emphasize surgical excision as the only therapeutic approach proven effective in treating dermatofibrosarcoma protuberans, highlighting the challenges of covering the resulting tissue loss from sometimes extensive and mutilating excisions.

CASE DESCRIPTION:

It concerned a 13-year-old girl from a good socio-economic background, with no particular medical history, who presented for management of a skin tumor on the scalp.

The symptoms began 2 years earlier with the appearance of a painless cutaneous swelling in the left mid-parietal region. Over time, the swelling progressively increased in size, its clinical appearance changed, and a second, ipsilateral swelling appeared at the parieto-occipital junction a year later (figure 1). They caused mild pain when pressed, headaches, and cosmetic concerns, prompting her to seek specialized care in our department.

On physical examination, two ulcerated nodular masses were found on the scalp. The larger one, located in the left mid-parietal region, measured 6 cm x 4 cm, while the smaller one, in the left parieto-occipital region, measured 3 cm x 1.5 cm.
The masses were oval-shaped, crusty in appearance, soft and cystic in consistency, with regular, well-defined borders. They were pruritic and painless to palpation, mobile relative to the deeper tissues, and showed local inflammatory signs such as redness and warmth.
Examination of the lymph node areas was unremarkable. The remainder of the clinical examination revealed no other abnormalities.

The brain CT scan, with and without contrast injection, revealed an isodense subcutaneous tissue image in the left mid-parietal and left parieto-occipital regions. There was no associated osteolysis of the underlying bone, and the mass moderately took up the contrast. No intracranial extension was observed.
The preoperative assessment was unremarkable.

Under general anesthesia, an excision biopsy was performed, removing the tumor masses as a single block (Figure 3), followed by histopathological examination and additional immunohistochemistry.

The morphological appearance and immunohistochemical profile favor a diagnosis of dual lesions of dermatofibrosarcoma protuberans, consistent with:
• Positive staining of tumor cells with anti-CD34 antibody.
• Negative staining of tumor cells with antibodies: Ps100, CD99, SMA.
• Ki67 proliferation index estimated at 40%.
• The posterior margin is involved by the lesion.

Therefore, the diagnosis of dermatofibrosarcoma protuberans is confirmed, and the patient is scheduled for further surgical excision with 3-4 cm oncological margins laterally and extending to a healthy anatomical barrier (periosteum) in depth (Figure 4 and 5).

Initially, the skin defect was left for natural healing to ensure histologically complete and adequate excision and to allow the patient a period of emotional recovery and rest. The wound showed favorable progress with almost complete epithelialization and reduction in size due to edge contraction mediated by myofibroblasts. However, this resulted in an unacceptable alopecic scar for a young girl (Figure 6).

Hence the interest in placing rectangular skin expansion prostheses until sufficient skin laxity is achieved (Figure 7) for the placement of an Orticochea flap (Figure 8).

The aesthetic and functional outcome is considered satisfactory by both the surgeon and the patient (Figure 9).

No signs of recurrence observed two years post-surgery.

DISCUSSION:

Dermatofibrosarcoma is an intermediate-grade dermal mesenchymal tumor [1]. It is a rare but not exceptional tumor, representing between 0.1% and 1% of malignant skin tumors. Taylor first described it in 1890 as a sarcomatous tumor resembling a keloid scar. Subsequently, dermatofibrosarcoma was reported in 1921 by Kuznitzky and Grabish, and then by Darier and Ferrand in 1924 [2], and Hoffman in 1925 [2], who named it "dermatofibrosarcoma protuberans". Pack and Tabah published the first significant series in 1951, and Taylor and Helwig established the histological characteristics of this tumor in 1962 [3].

Dermatofibrosarcoma protuberans, as described by Darier and Ferrand, can occur at any age, with average ages at diagnosis ranging from 28 to 47 years [1]. As described by several authors, this tumor is rare in children under 15 years old, and congenital forms are extremely rare [4].

Our patient is notable for her very young age (13 years), as well as the location of the lesions. Typically, dermatofibrosarcoma protuberans, as reported in the literature, occurs in 50 to 60% of cases on the trunk and abdomen, in 20 to 30% of cases at the root of the limbs, and only 15 to 20% are attributed to the head and neck [5]. The palms of the hands and soles of the feet are usually spared according to some authors.

As for the initial lesion, it typically starts as a small, pinkish fibrous plaque or a hard nodule embedded in the skin, which gradually expands to form a tumor-like lesion: painless, brown or pink in color, and sometimes telangiectatic. These nodules may occasionally ulcerate [6].

In rare cases like ours, due to delayed consultation, the tumor presents as a multinodular dermo-hypodermal plaque consisting of multiple nodular, bossed, confluent, hard, and mobile skin tumors. It forms a large, papulokeratotic plaque that is crusty and pruritic, giving the skin a dirty, rough texture upon palpation (Figure 1).

On average, the tumor measures between 1 to 5 cm in size. According to reports, cases of tumors described as "monstrous" have been documented with weights reaching up to 6.5 to 7 kg [7]. In our patient, the largest tumor measured 6 cm x 4 cm, while the smaller one measured 3 cm x 1.5 cm.

In 10 to 20% of cases, there is a history of local trauma preceding the onset of dermatofibrosarcoma protuberans. According to a series by Taylor and Helwig, including 115 cases of dermatofibrosarcoma protuberans, a history of trauma was found in 16.5% of cases [8]. It is noteworthy that our patient did not report any history of local trauma.

Histological examination is essential to confirm the diagnosis of dermatofibrosarcoma protuberans. It reveals a dense cellular proliferation occupying the dermis entirely, often poorly defined and non-encapsulated. It extends with thin projections, sometimes deeply into the subcutaneous tissue (Figure 2a), explaining the occurrence of recurrences even with wide resection margins. The epidermis remains intact. The cells are spindle-shaped, elongated, with oval, regular nuclei and variably abundant cytoplasm. Mitoses are variable, with rare atypia. The stroma varies from one area to another [9].

On the architectural level, the cells are arranged in radial bundles with a swirling pattern often described as "wheel spoke" appearance. Necrotic areas are rarely encountered [10].

In our case, there was a fairly well-defined tumor proliferation, densely cellular, composed of bundles of spindle-shaped cells, occasionally adopting a storiform pattern. The nuclei were elongated, irregular, sometimes with mitotic activity. The cytoplasm was scant, eosinophilic, and pale. There was no tumor necrosis present.

The histological appearance guides the diagnosis in the majority of cases, but occasionally, there may be cases that are ambiguous and confusing. This highlights the importance of immunohistochemistry, which helps distinguish dermatofibrosarcoma protuberans from other spindle cell tumors. Immunohistochemical staining typically shows intense and diffuse positivity for CD34 (Figure 2b), focal positivity for smooth muscle actin

(SMA), and consistent negativity for S100 protein and desmin [11]. Areas of sarcomatous transformation rarely express CD34 and very weakly [12].

In our patient, the immunohistochemical panel showed:
- Positive staining of tumor cells for CD34 antibody.
- Negative staining of tumor cells for antibodies: S100, CD99, smooth muscle actin (SMA).
- Ki67 proliferation index estimated at 40%.

Regarding cytogenetic analysis, it reveals the presence of supernumerary ring chromosomes, formed from sequences derived from chromosomes 17 and 22, or more rarely from t (17;22) translocations [13].

The CT scan assessment allows for evaluating the depth extension of tumors and ruling out potential bone lysis or any endocranial expansive process.

The treatment of dermatofibrosarcoma protuberans is challenging due to its potential for subclinical extension, which explains its high recurrence rate. Wide and extensive surgical excision is the standard treatment, with recommended lateral margins of 4 to 5 cm and deep removal of the superficial fascia [14]. Some advocate for postoperative radiotherapy after the second recurrence. Systemic chemotherapy is no longer recommended [15].

The slow progression and high recurrence rate of this tumor necessitate strict and rigorous clinical surveillance.

The repair of the tissue loss caused by excision involves all possible methods of plastic surgery, ranging from direct suturing if feasible without excessive tension, to directed healing and dermo-epidermal grafting if the vascularization of the recipient site allows. Skin and musculocutaneous flaps are used, especially for aesthetic reasons (scalp, breast...), though they do not facilitate as thorough surveillance for detecting potential recurrences as grafts do.

Regarding our patient, an excision biopsy was initially performed under general anesthesia, removing the tumor mass in one piece (Figure 3), followed by orientation of the surgical specimen for pathological examination.

Additional immunohistochemistry study was necessary to confirm the diagnosis of dermatofibrosarcoma protuberans. The findings indicated posterior and deep lesion margins, prompting a second surgical procedure for further excision with safety margins of 3 - 4 cm laterally and down to the periosteum in depth (Figures 4 and 5).

The surgical specimen was oriented and sent to pathology to ensure the completeness and adequacy of the excision.

The size of the induced tissue defect was reduced by approximating the edges using Blair-Donati sutures.

The coverage of the induced tissue loss occurred secondarily, following absolute histological confirmation that the excision was sufficiently complete.

Direct suturing was not feasible due to the size of the tissue defect (15 cm x 9 cm). Directed healing was possible, as was dermo-epidermal grafting, given adequate vascularization of the recipient site (because the periosteum was preserved). However, these options carried the risk of an alopecic scar, which is unacceptable in a young female patient.

For aesthetic reasons, coverage with an Orticocchea flap, following skin expansion using tissue expanders, was the best choice for this patient. The procedure was performed in two stages:

In the first stage, two rectangular tissue expanders of 100 cc volume each were placed: one on the right temporo-parietal area and the other on the left temporal area. Multiple filling sessions with saline solution, at a rate of 20 cc per week, were conducted until achieving the necessary skin expansion (Figure 7).

In the second stage, an Orticocchea flap was raised, utilizing the remaining scalp tissue. The flap was elevated extensively and fragmented into several segments, ideally based on a pedicle. This technique is advantageous for closing extensive tissue defects while minimizing the alopecic area (Figure 8).

Two postoperative complications occurred in our patient: a subcutaneous fluid collection treated with repeated needle aspirations, and superficial necrosis of some edges managed with directed healing after chemical debridement, without significant scarring consequences.

The aesthetic and functional outcome has been deemed satisfactory by both the patient and the surgeon after 7 months post-operation (Figure 9).
Postoperative radiotherapy was not indicated for our patient.

The prognosis is primarily determined by the local malignancy of this tumor [14]. Darier and Ferrand's dermatofibrosarcoma protuberans almost never metastasizes, and lymph node involvement occurs in less than 1% of cases [16]. In our patient, no local or distant lymph node extension was found.

It is a clinically challenging entity due to its high recurrence rate, even with extensive surgical excision. According to the literature, local recurrence occurs in 20 to 40% of cases [1]. Death is rare and typically occurs late due to local complications [17]. In our case, there has been no local recurrence or metastasis observed during the first two years following surgical excision.

CONCLUSION:

Dermatofibrosarcoma protuberans is a rare fibrous tumor of the skin, characterized by diagnostic challenges, indolent local growth, and a high potential for local recurrence, necessitating extensive surgical excision with histological control of the surgical specimen.

The reconstruction of the tissue defect resulting from excision should only be performed secondarily and poses a significant challenge for the plastic surgeon, especially in specific anatomical locations. It must meet two main objectives: covering the tissue loss and restoring the best possible functional and aesthetic outcome.

PATIENT CONSENT

the patient's parents provided informed consent prior to his involvement in the study, acknowledging his understanding of the research procedures, risks, and confidentiality.

ETHICAL APPROVAL:

Ethical approval is considered unnecessary by the ethics committee of the hospital, because this is a unique case encountered during practice and which does not involve any experimentation on humans or animals.

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GUARANTOR:

All authors of the article have approved its publication and supervised its writing.

DECLARATION OF COMPETING INTEREST:

The authors have no conflicts of interest or financial disclosures.

AUTHOR CONTRIBUTION

A. MESFIOUI (Corresponding author): study concept, data collection, writing the paper.
M.A HAOUANE, M.A AZAMI: Histological Study and Histological Imagery.
F.Z. NAFIDI, A. ARROB, T. NASSIM SABAH: supervision, review & editing.
K. TOURABI: writing the paper, reviewing and validating the manuscript's credibility

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THE FIGURES

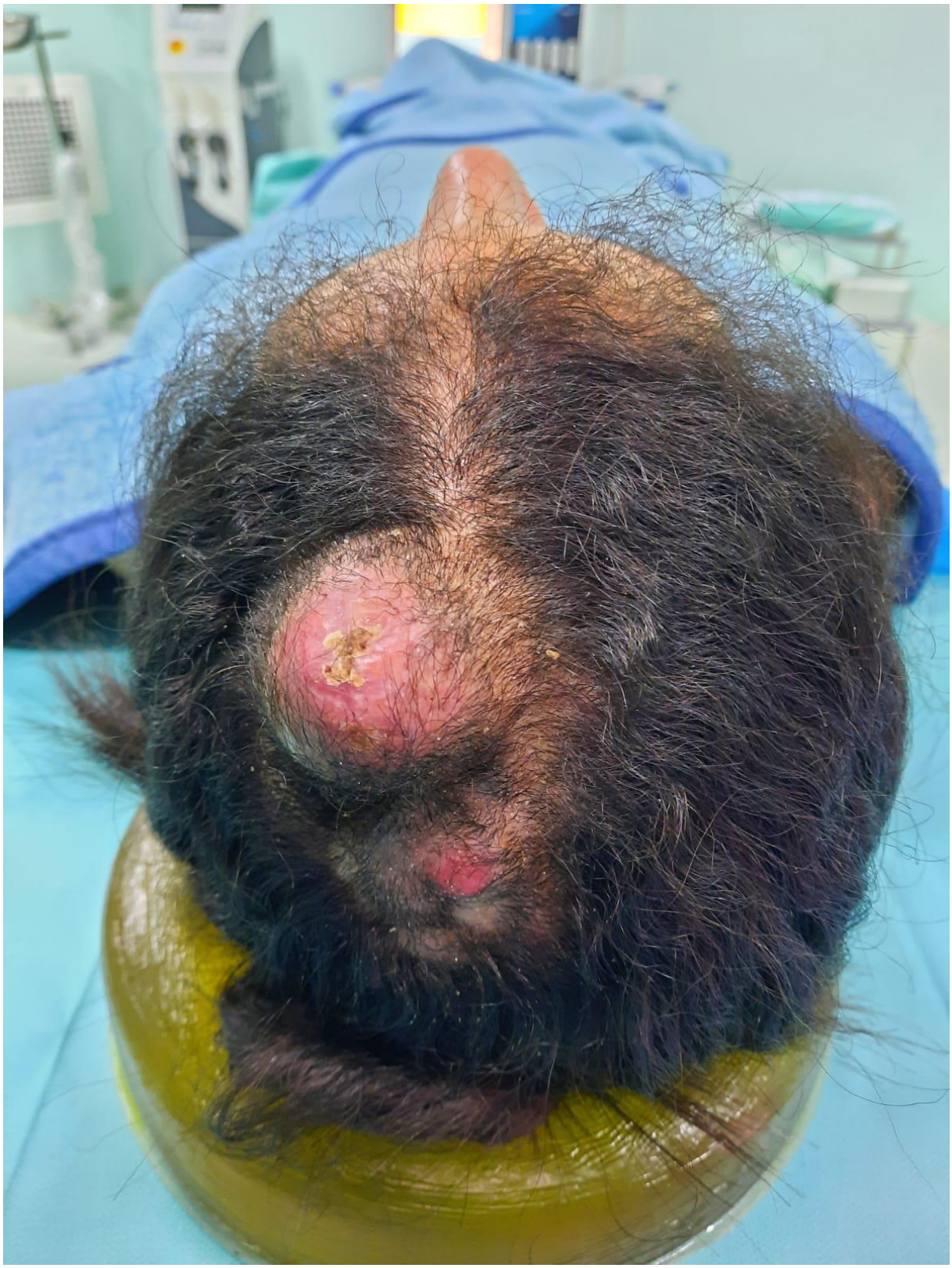

Figure 1: Clinical appearance of a scalp dermatofibrosarcoma protuberans in a 13-year-old girl.

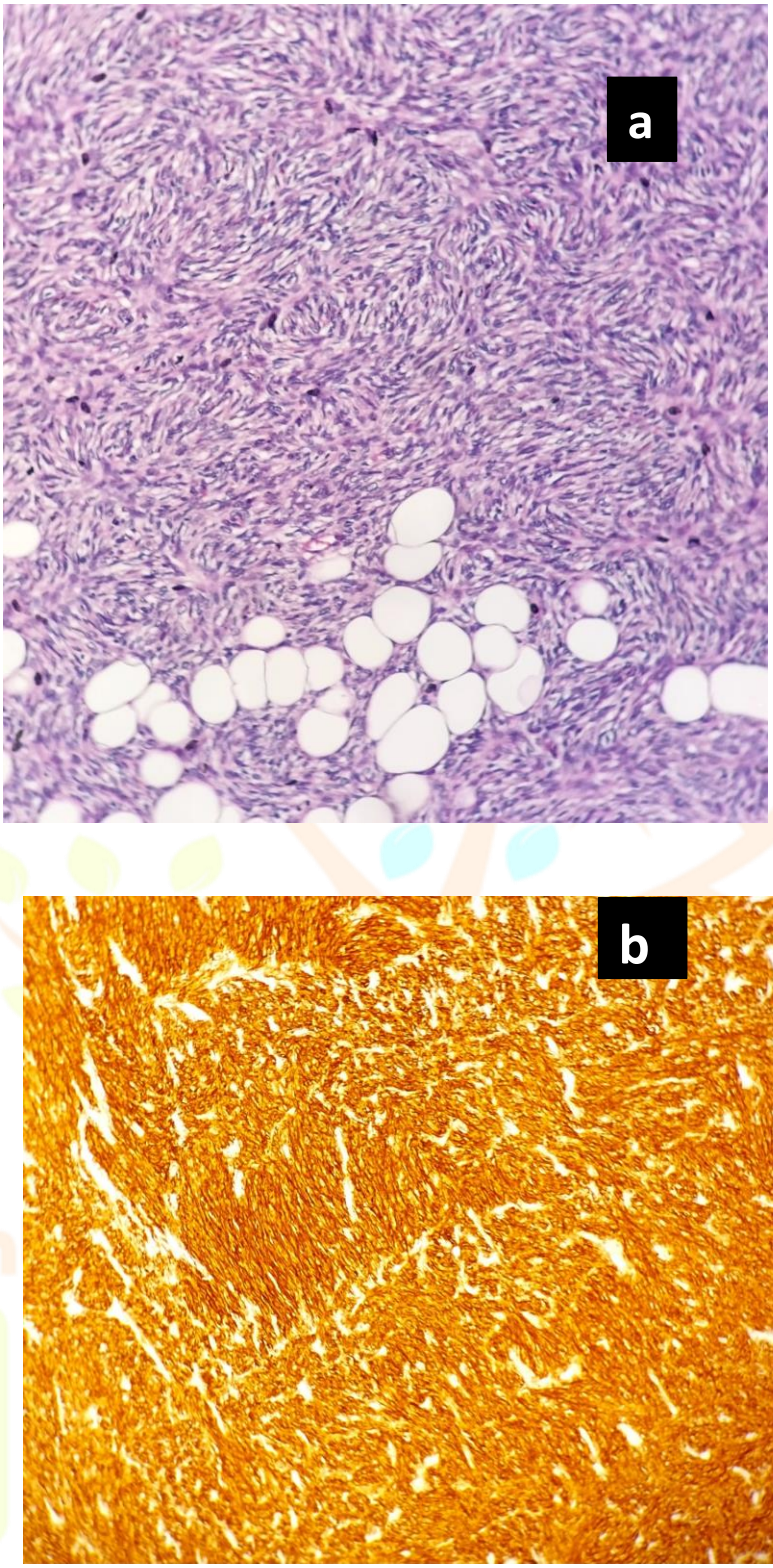

Figure 2: The histological study with immunohistochemical complement of a dermatofibrosarcoma protuberans:
a - Higher magnification hemateine-eosine slide showing Spindle cells with characteristic storiform architecture and infiltration of subcutaneous adipose tissue with a so-called honeycomb pattern.
b - Immunohistochemistry for CD34 showing diffuse staining.

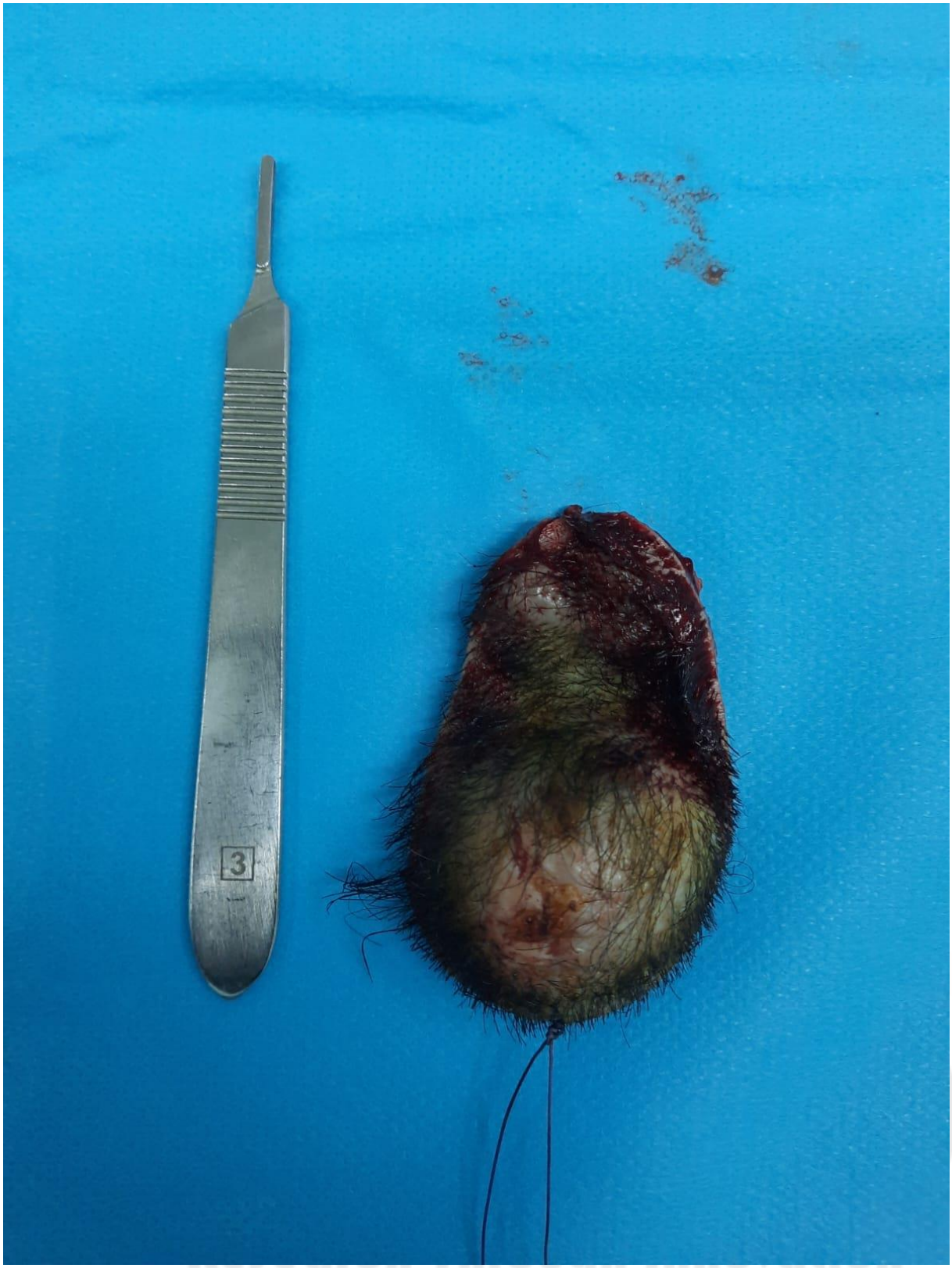

Figure 3: Surgical specimen of biopsy-excision of a scalp dermatofibrosarcoma protuberans, oriented with two anterior sutures towards the forehead.

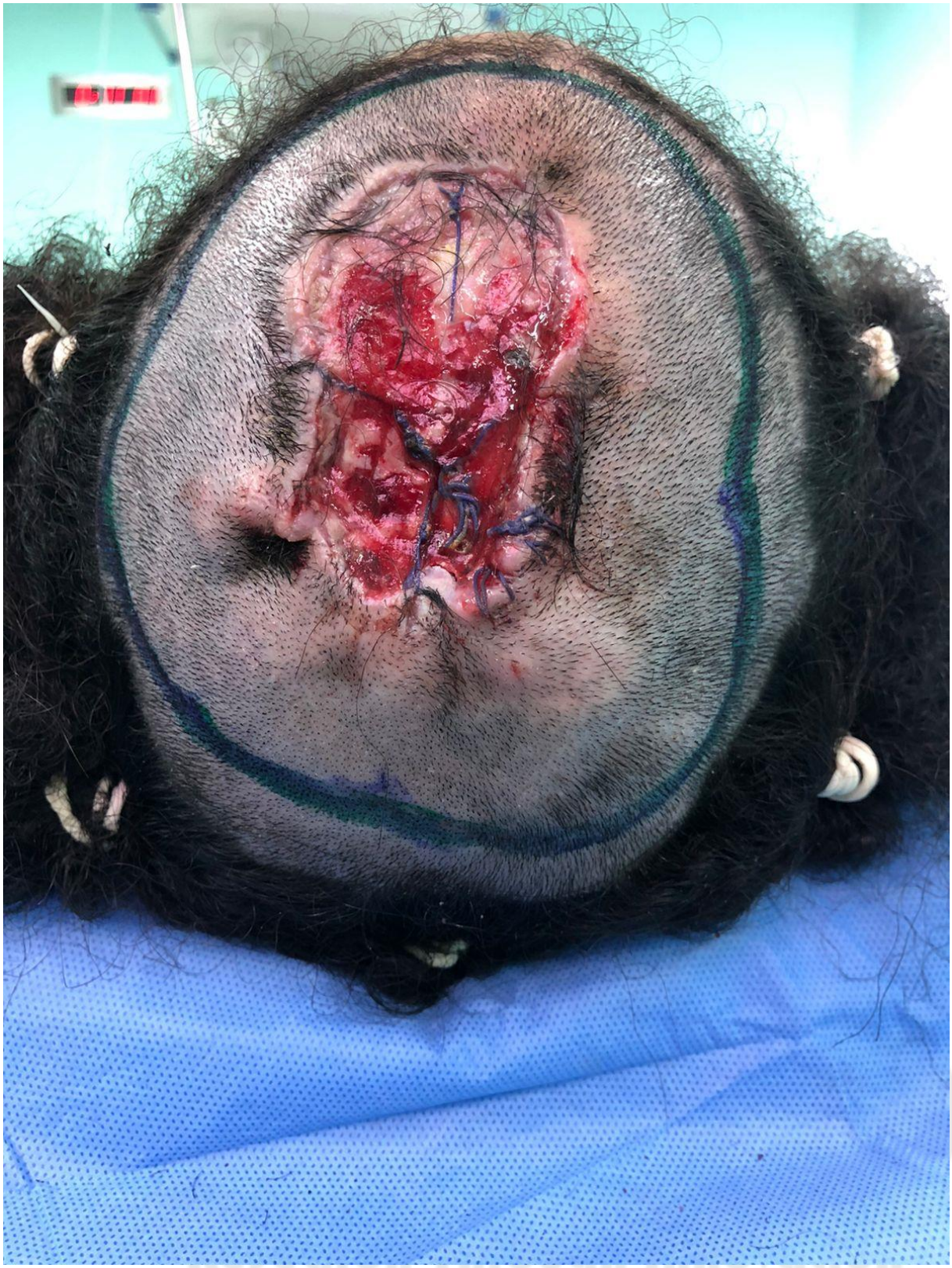

Figure 4: Preoperative surgical outline for additional excision of scalp dermatofibrosarcoma protuberans with lateral safety margins of 3 to 4 cm.

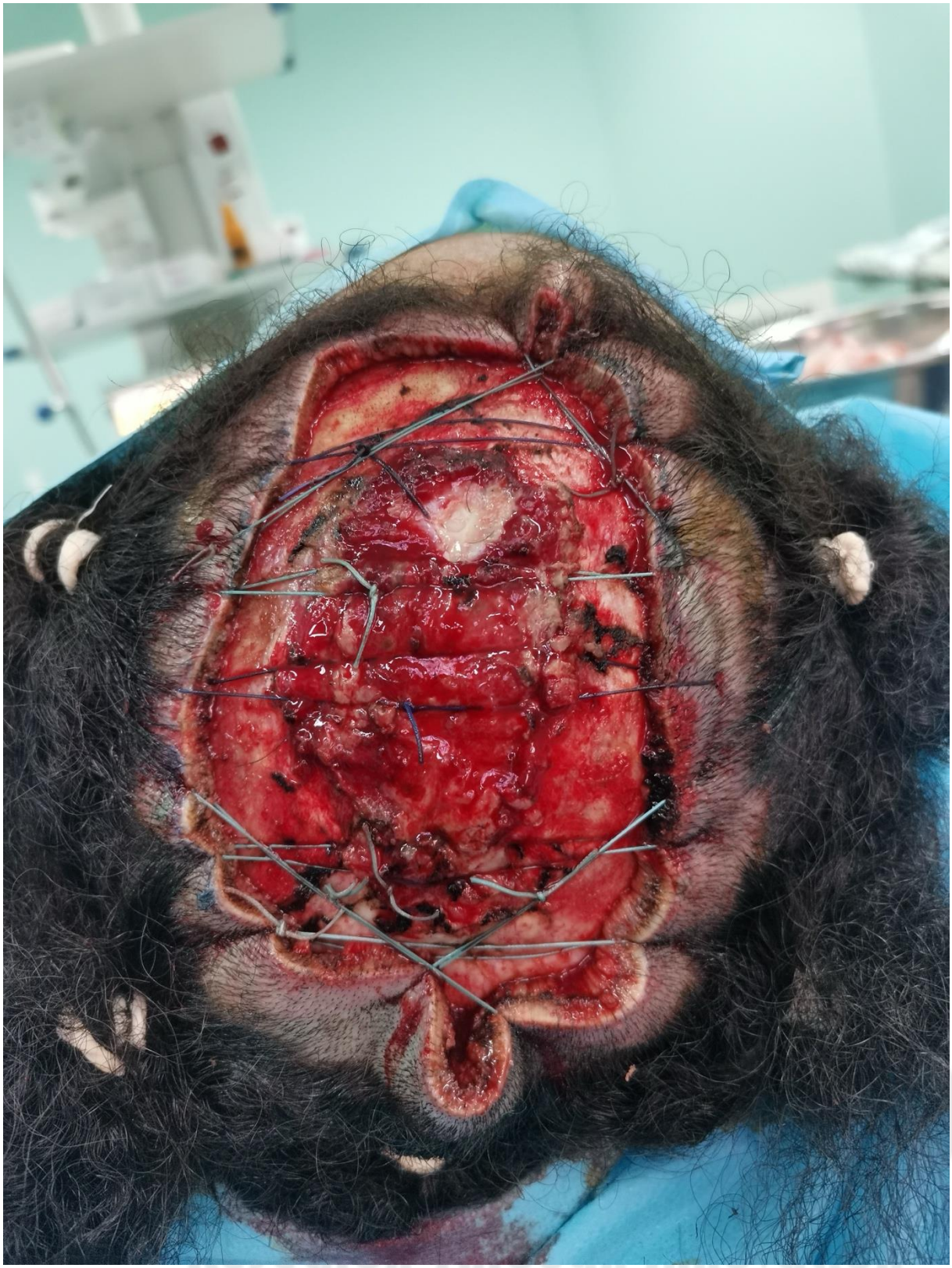

Figure 5: Immediate postoperative appearance of the tissue defect following additional excision of scalp dermatofibrosarcoma protuberans.

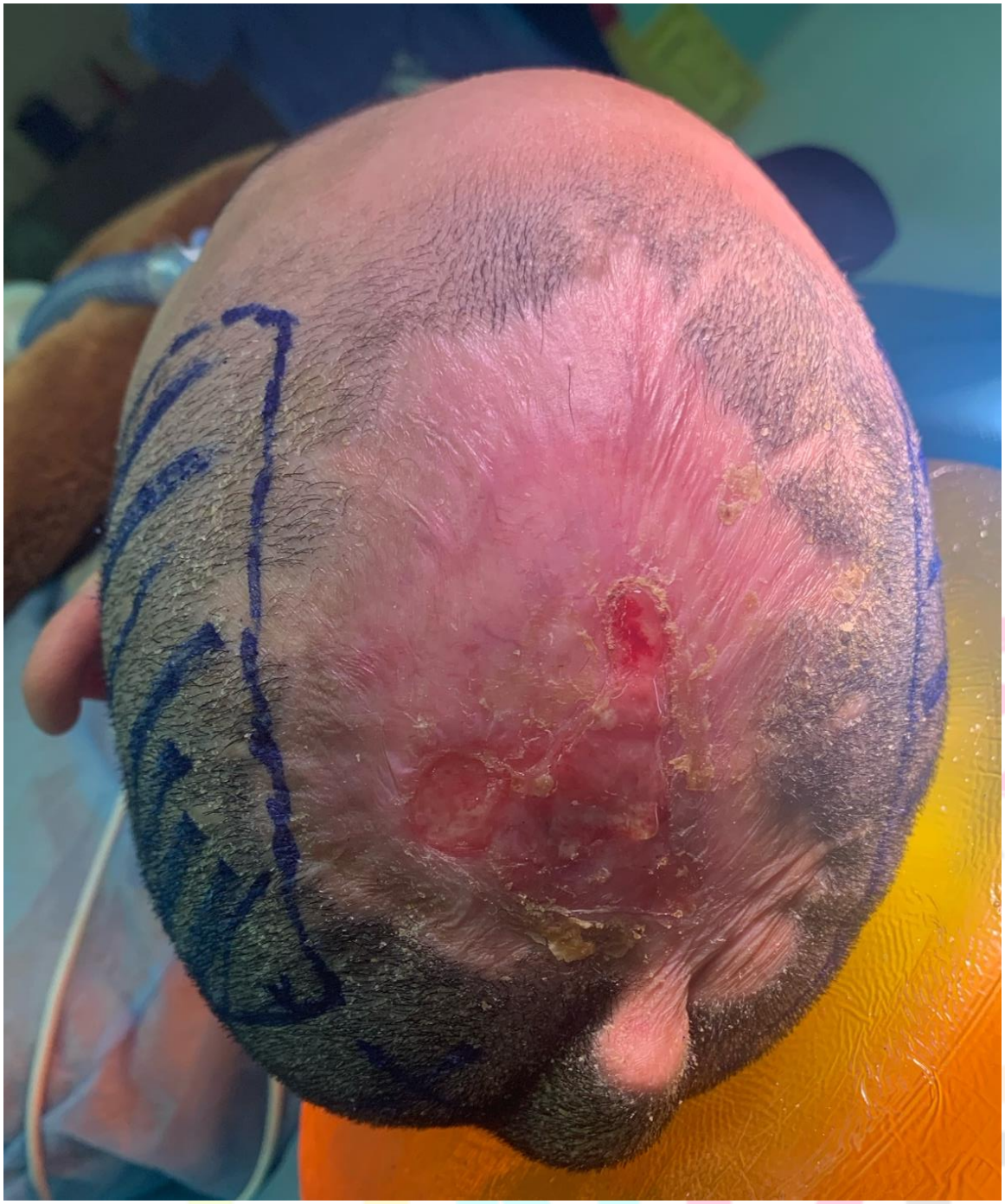

Figure 6: Appearance of the tissue defect undergoing directed healing, 5 months post excision of scalp dermatofibrosarcoma protuberans.

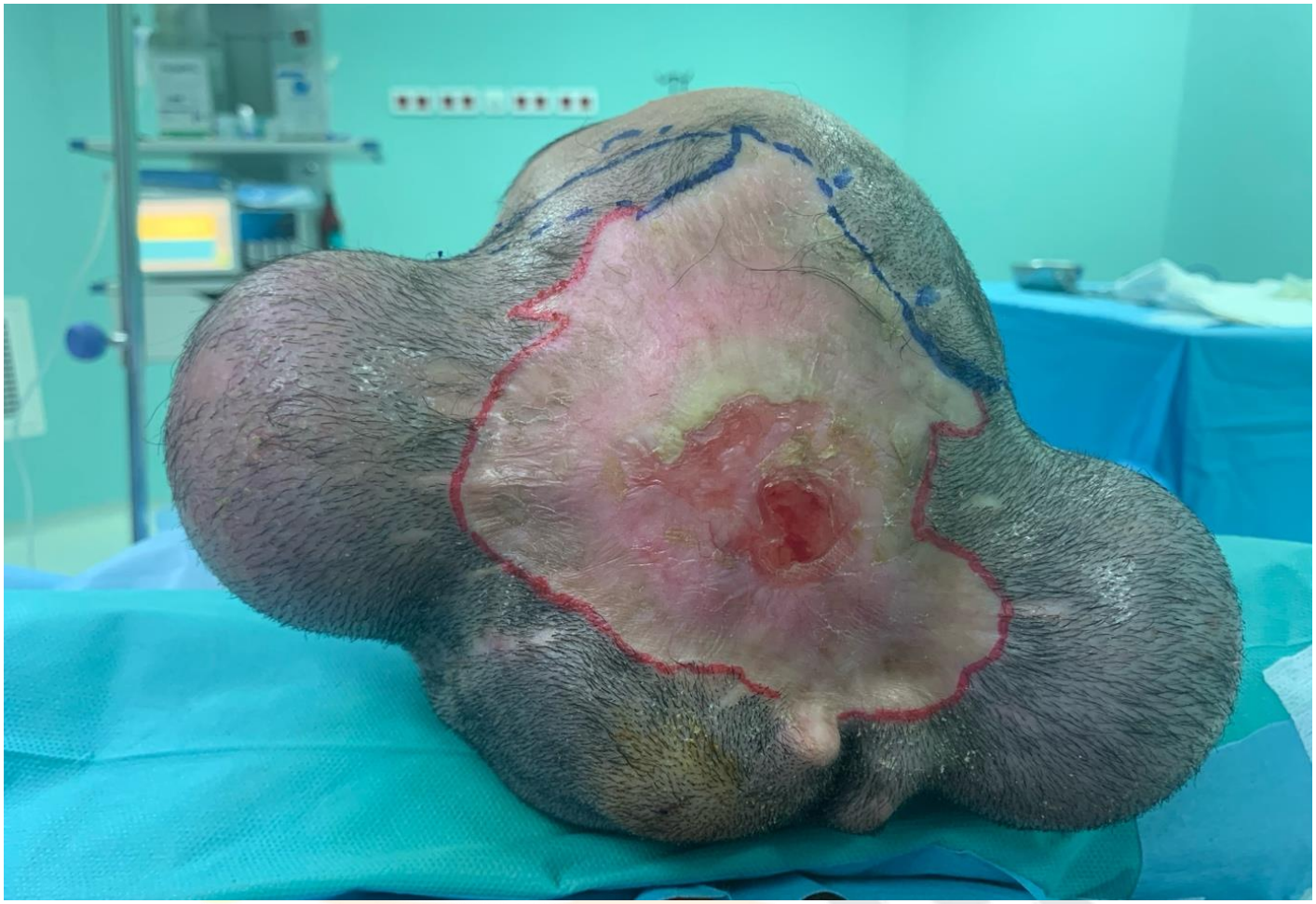

Figure 7: Scalp skin expansion using rectangular tissue expanders (appearance at the end of filling).


a

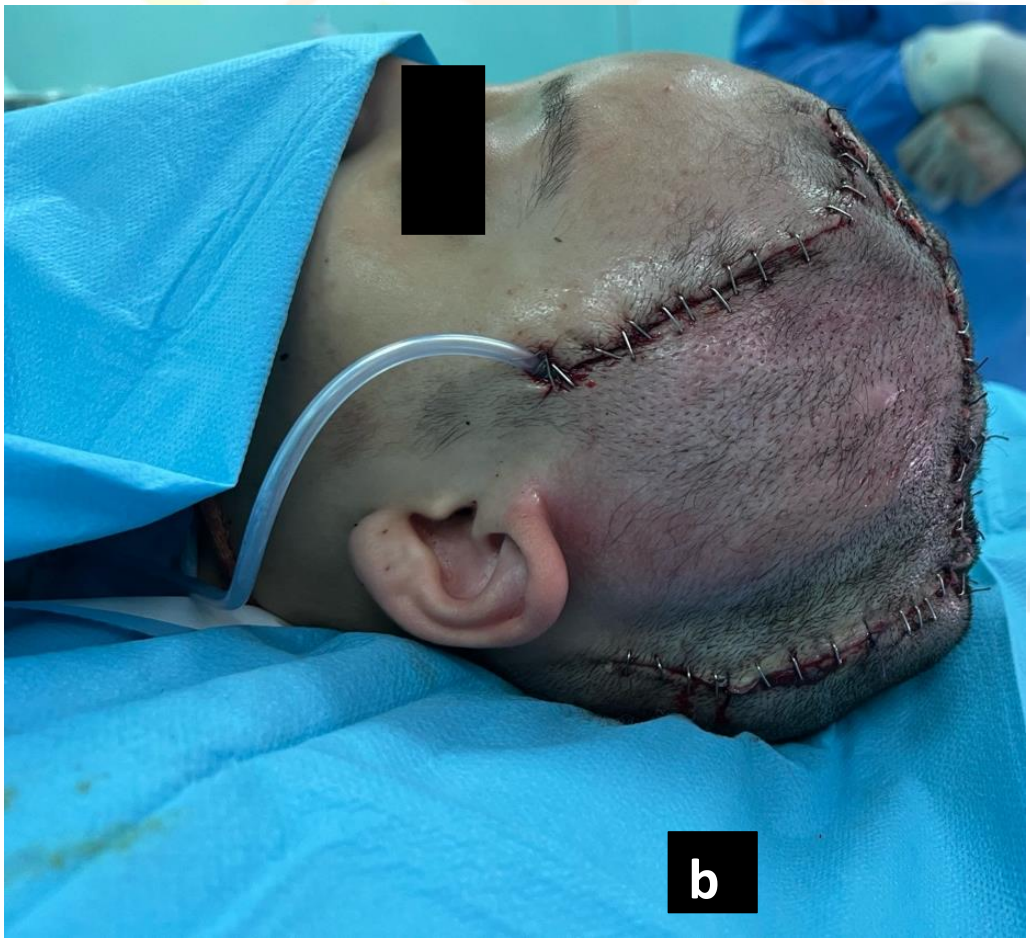
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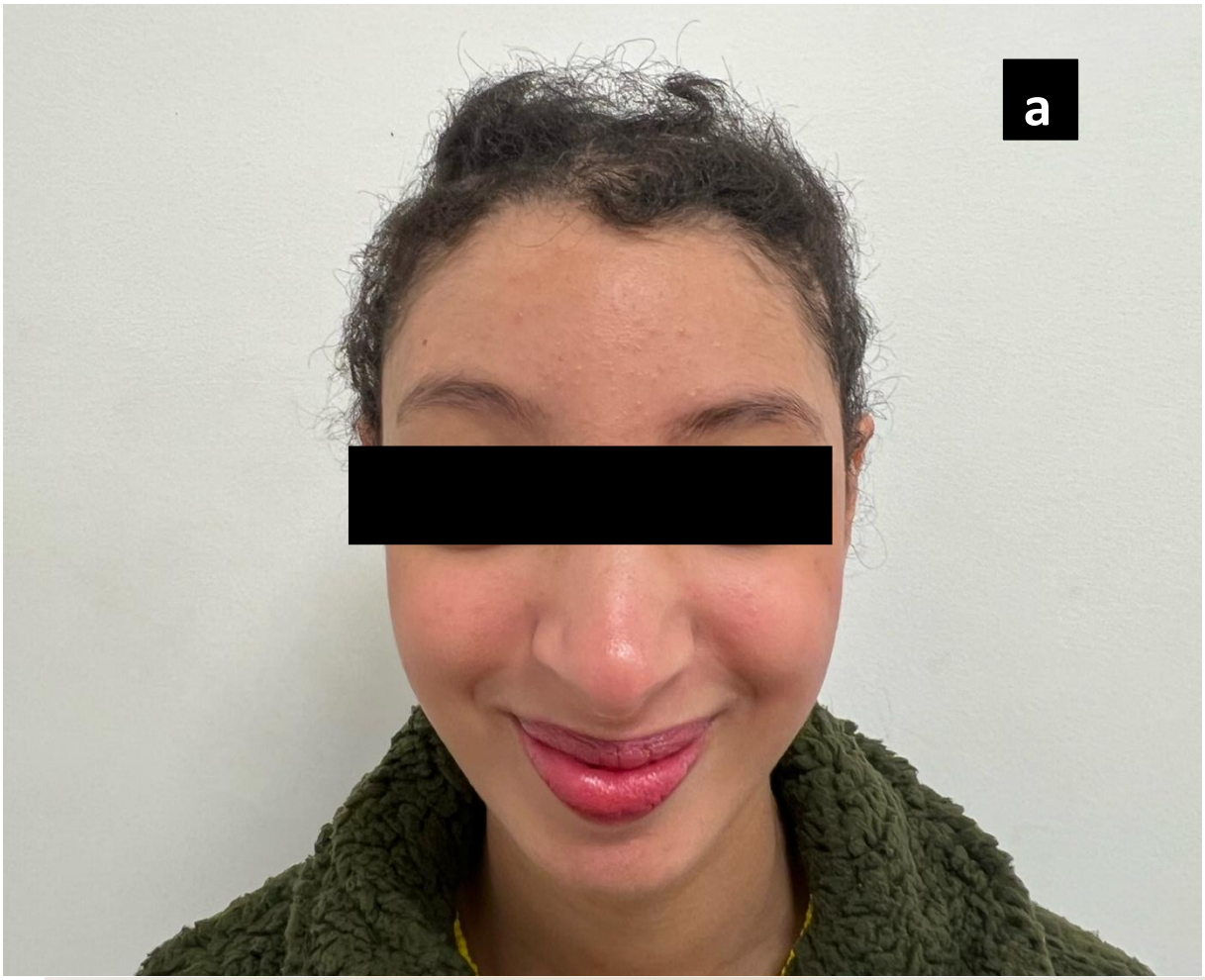
Figure 8: Immediate postoperative appearance of coverage of the defect induced by excision of scalp dermatofibrosarcoma protuberans using an ORTICOCHEA flap after removal of skin expansion prostheses.
a. Superior view of the scalp
b. Left lateral view of the scalp
c. Right lateral view of the scalp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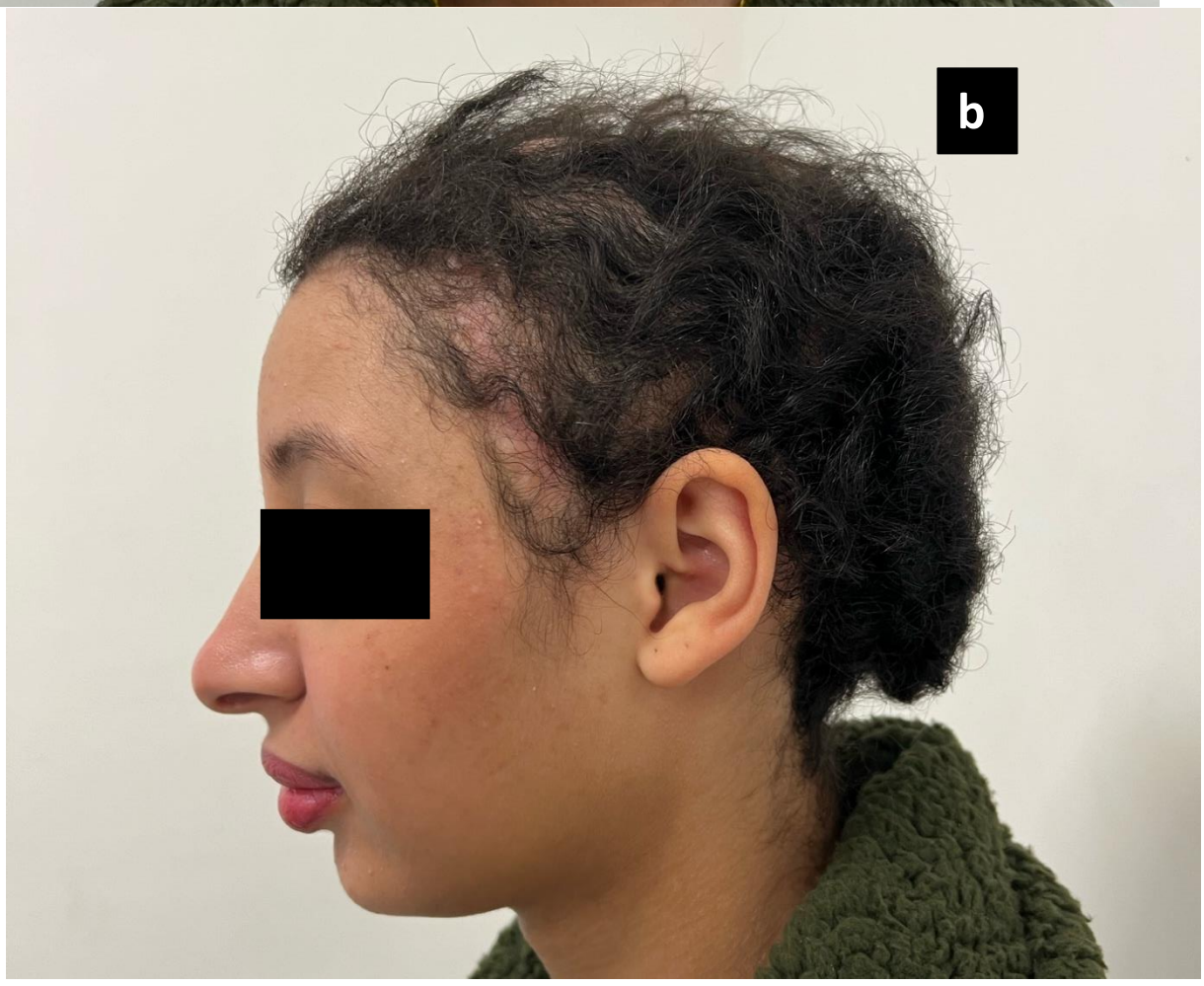
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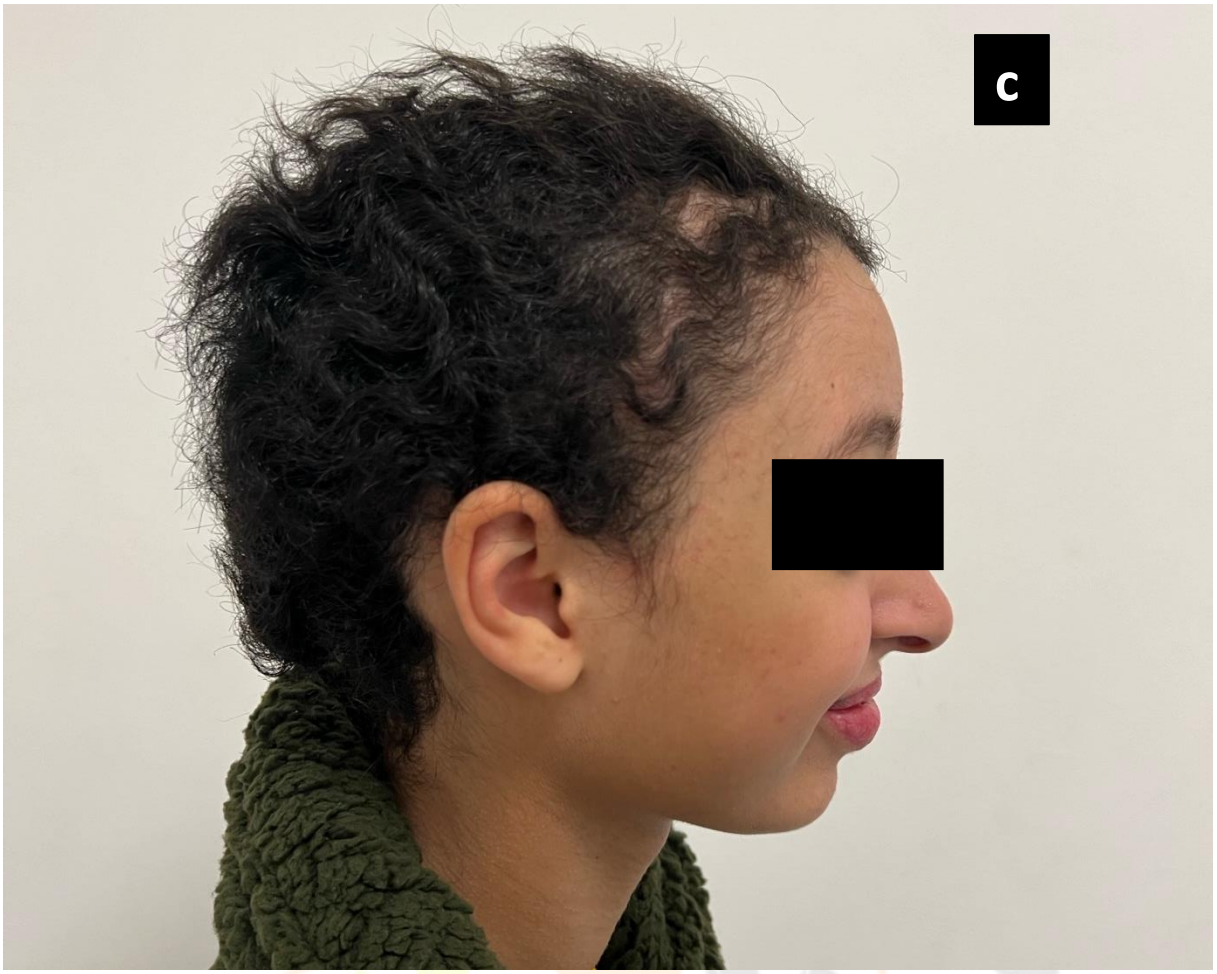

Figure 9: 7-month postoperative result of reconstruction of a defect induced by excision of scalp dermatofibrosarcoma protuberans using an ORTICOCHEA flap.
a. Frontal view
b. Left profile view
c. Right profile view